

Case Report

A Case of Retroperitoneal Desmoid-Type Fibromatosis that Invaded Skeletal Muscle after Left Hepatectomy and Cholecystectomy

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Abstract

Background: Desmoid Fibromatosis (DF) is clonal fibroblast hyperplasia that occurs in deep soft tissue and is a type of fibroblast tumor with local invasion potential.

Case presentation: A 52-year-old woman visited a hospital and a physical examination revealed retroperitoneal mass that had been present for more than 1 month. Three years prior, left hemangioma of the liver was treated with left hepatectomy and cholecystectomy. The physical examination revealed no abdominal pain, abdominal distension, acid reflux heartburn, vomiting or other symptoms. The results of the laboratory examination were as follows: blood potassium concentration (3.01 mmol/L), blood magnesium concentration (0.54 mmol/L), and urinary calcium concentration (0.34 mmol/24h). Routine blood work, tumor markers, and blood coagulation test results were generally normal.

Conclusion: The possibility of retroperitoneal DF should be considered when clinicians find isointensity on T1W imaging, hyperintensity on T2W imaging and progressive enhancement on MRI after peritoneal tumor, especially in patients with a surgical history.

Keywords: Desmoids - type fibromatosis; Retroperitoneal; Case report

Abbreviations

DF: Desmoid Fibromatosis; SMA: Smooth Muscle Actin; FSCE-T1W: Fat Suppression Contrast Enhanced T1 Weighted

Introduction

DF is a rare mesenchymal neoplasm that results from the proliferation of fibroblasts or myofibroblast-like tumor cells as shown by histology. Although DF is histologically benign, it is locally invasive and frequently recurs in 39% to 79% of the cases [1], however, it does not metastasize [2,3]. DF is a rare disease and accounts for 0.03% of tumors and 3% of all soft tissue tumors [4,5]. Moreover, DF may arise throughout the body, and is clinicopathologically classified into three types: extra-abdominal (60%), such as trunk, head, neck and limbs; abdominal wall (25%), which often occurs in the abdominal wall muscles and intra-abdominal (8% to 15%), such as mesentery and retro peritoneum. The number of tumors that occurs within the peritoneum is very rare, and retroperitoneal DF accounts for less than 1% of retro peritoneal masses [6], varying from 0.6% to 4% of all sites of desmoids tumors [7,8]. Herein, we report a case of DF that

occurred in the retroperitoneal space and invaded skeletal muscle after left hepatectomy and cholecystectomy.

Case Presentation

A 52-year-old woman visited a hospital and a physical examination revealed retroperitoneal mass that had been present for more than 1 month. Three years prior, left hemangioma of the liver was treated with left hepatectomy and cholecystectomy. The physical examination revealed no abdominal pain, abdominal distension, acid reflux heartburn, vomiting or other symptoms. The results of the laboratory examination were as follows: blood potassium concentration (3.01 mmol/L), blood magnesium concentration (0.54 mmol/L), and urinary calcium concentration (0.34 mmol/24h). Routine blood work, tumor markers, and blood coagulation test results were generally normal. Abdominal magnetic resonance imaging showed a 4.3 cm × 3.3 cm mass in the right retroperitoneal space and after the head of the pancreas. The mass was isointense compared to muscle on T1-weighted imaging (Figure A1), inhomogeneously intense, but hyperintense compared to muscle on T2-weighted imaging (Figure A2), and slightly hyperintense on diffusion-weighted imaging (Figure A3). The tumor showed contrast enhancement and progressive delayed heterogeneous reinforcement (Figures A4 and 5). We also can find the mass was closely connected to the inferior vena cava, portal vein, common hepatic artery and abdominal trunk (Figure A6). Based on these imaging signs, a malignant tumor was suspected. In response, we performed tumor resection, the original incision under the right costal margin was extended into the abdomen, where it could contact retroperitoneal masses and enlarged lymph nodes after pancreatic head. The mass was approximately 5 cm × 4 cm in size, and had a hard quality and poor movement. It was attached to

Citation: Wang X, Lei li, Sun X. A Case of Retroperitoneal Desmoid-Type Fibromatosis that Invaded Skeletal Muscle after Left Hepatectomy and Cholecystectomy. *Ann Clin Case Stud.* 2020; 2(4): 1034.

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Publisher Name: Medtext Publications LLC

Manuscript compiled: Sep 13th, 2020

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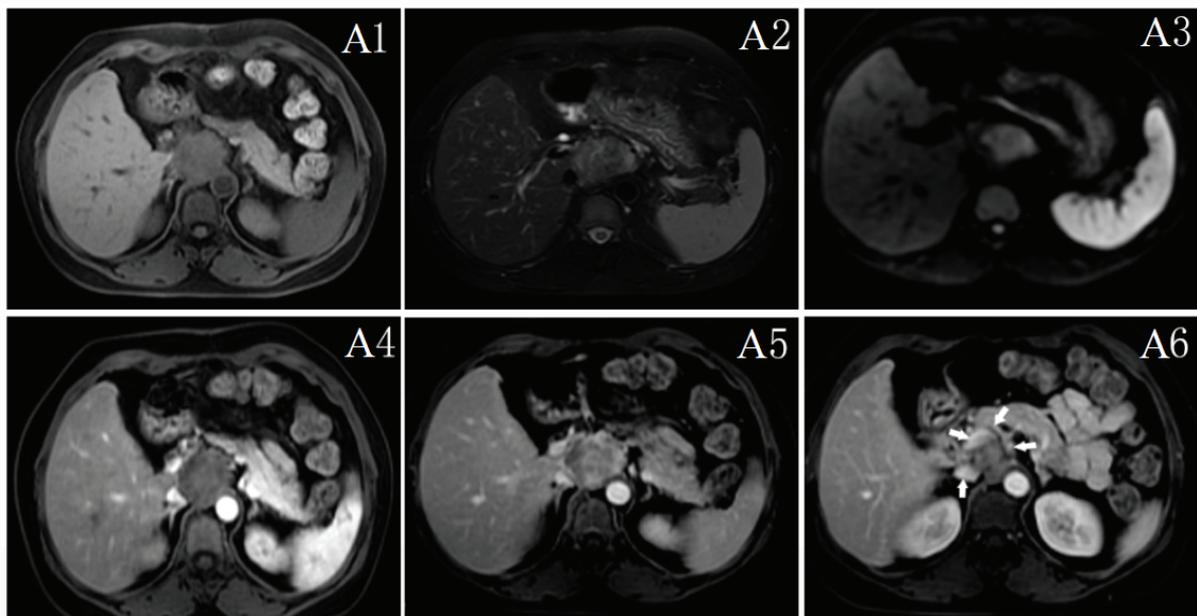


Figure A1: T1-weighted image; A2: T2-weighted image; A3: Diffusion-weighted image; A4: Early-phase image; A5: Late-phase image. Abdominal magnetic resonance imaging showed a right retroperitoneal tumor, which were isointense compared to skeletal muscle on T1-weighted imaging, hyperintense compared to skeletal muscle on T2-weighted imaging and slightly hyperintense on diffusion-weighted imaging. The tumor showed contrast enhancement and progressive delayed heterogeneous reinforcement. The mass was closely connected to the inferior vena cava, portal vein, common hepatic artery and abdominal trunk (arrows).

the surrounding tissue and insert duodenum ascending serosa layer. Histopathologically, the tumor was composed of spindle-shaped fibroblast-like cells that infiltrated into the skeletal muscle and fatty tissues (Figure B1); however, atypical cells were not found in the tumor. Immunohistochemical analysis showed that tumor cells were partly positive for Smooth Muscle Act (SMA) in and β -catenin in (Figure B2), but negative for S-100 protein, CD34 and CD117, and that the Ki-67 labeling index was 1%. Based on these results, we diagnosed the tumor as DF. The postoperative recovery was good, and the patient was discharged on postoperative day 21. The patient remained asymptomatic during a 9-month follow up period.

Discussion

DF was first named by Mueller in 1938 and is a fibrous tumor formed by the clonal proliferation of fibroblasts or myofibroblasts of deep soft tissue (muscle, fascia, or aponeurosis) In 2013, WHO

classified this tumor as fibroblast/myofibroblast tumors, on the basis of its biological behavior, which was between that of benign and malignant tumors, and as intermediate (local invasiveness). Due to its invasiveness, it is also known as an invasive fibroma or a hard fibroma. The peak age of disease onset is 25 to 35 years old, and DF commonly occurs in women, particularly women of childbearing age. The male to female ratio is approximately 1.1:1.2 [9]. However, intra-abdominal fibromatosis shows no gender difference or age predilection [10]. This case is consistent with the clinical characteristics of intra-abdominal fibromatosis. The etiology of the disease is not known. A previous report stated that DF was associated with trauma, and surgical wounds, for example: right radical orchiectomy for a testicular germ cell tumor emergent explorative laparotomy for a ruptured GIST [11,12], cesarean section and total abdominal hysterectomy and left salpingo-oophorectomy due to uterine myoma and a paraophoritic

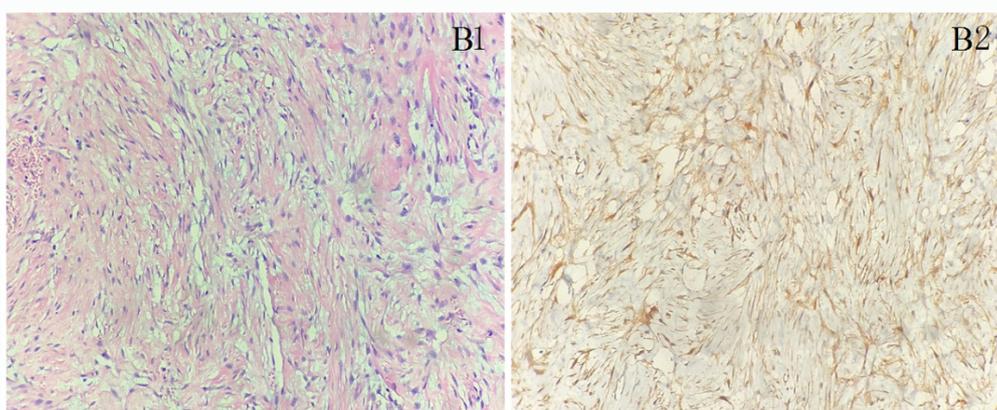


Figure B1: Microscopically, the tumor was composed of spindle-shaped fibroblast-like cells that infiltrated into the skeletal muscle, HE stain, $\times 200$; B2: Immunohistochemical analysis of the tumor shows positive results for β -catenin in spindle cells, HE stain, $\times 200$.

cyst [13], and Familial Adenomatous Polyposis (FAP) in Gardner syndrome, DF arises in approximately in 5% of cases of familial adenomatous polyposis [14]. In the present case, the patient had a history of left hepatectomy and cholecystectomy 3 years before detection of the retroperitoneal tumor, Although the etiology of DT in this case is unclear, the literature reports that 50% of these cases developed in the first 4 years post-trauma or surgery [15,16]. The present case of postoperative retroperitoneal desmoids occurred within an appropriate latent period. Most patients present with an asymptomatic abdominal mass, as in this case, but some have symptoms related to compression and invasion of adjacent organs, such as invasion of the ureter causing obstruction [13], occurrence in the pelvic cavity causing obstructed labor [17], and occurrence in the right anterior sub hepatic space causing acute duodenal obstruction [18], abdominal pain occurs in all of the above cases.

The MRI appearance of DT depends on the relative level of fibroblast proliferation. According to Yong-Woo et al. [19], tumors had iso-Signal Intensity (SI) relative to muscle on T1W images, low SI on T2W images, and relatively homogeneous strong enhancement on Fat Suppression, Contrast-Enhanced T1-Weighted (FSCE-T1W) images. Moreover, MR images indicate compression of the inferior vena cava and encasement of the right common iliac artery. According to the presentation described by Hitomi et al. [13], tumors were isointense compared to skeletal muscle on T1 and T2 weighted imaging and slightly hyperintense on DWI. Furthermore, tumors showed contrast enhancement, which was slight in the early phase and strong in the late phase and MRI showed tumor involvement in the unilateral ureter. Our findings are similar to those of previous reports on T1-weighted and diffusion-weighted imaging and enhancement, but T2-weighted imaging was different from previous articles. This discrepancy may be because the levels of fibroblast proliferations in DT are different, and when there are more fibers, the T2 signal is lower. The most common retroperitoneal neoplasm is lipogenic, but lipogenic neoplasm's can often be identified because they contain adipose components. Therefore the main differential diagnoses are Castleman disease or lymphoma of the retro peritoneum, neurogenic tumors, and leiomyosarcoma. The differential diagnosis between Castleman disease and lymphoma and DF are DWI signal and enhancement. In Castleman disease or lymphoma of the retroperitoneum due to the tight arrangement of cells, the diffusion of cells is limited; leadings to clearly high DWI-signal and showings obvious reinforcement. In view of this feature, the two diseases can be distinguished. In this case, the enhancement method needs to differentiate between the paragangliomas and schwannomas in retroperitoneal neurogenic tumors. Neurogenic tumors can form along the retroperitoneal nerves, but paragangliomas and schwannomas are prone to cystic degeneration and necrosis. On MR, the differential diagnosis between leiomyosarcoma and this disease is difficult. Leiomyosarcoma has the highest cystic change and necrosis rate among sarcomas, and is prone to infiltrating growth, often accumulating retroperitoneal blood vessels, which may be identified by these indirect signs, but ultimately requires pathological diagnosis.

The best treatment for DF is surgical resection, but postoperatively DF has a high recurrence rate, the 10 year recurrence rate of desmoid-type fibromatosis was reported to be 33%, and more than 90% of recurrences occurred within 5 years. Thus, reducing the recurrence rate is more important. Panagiotis et al. [20], reported that clear resection margins had a significantly lower risk of local recurrence compared to an intralesional margin, but Broekhoven et al. [21], failed

to confirm this relationship. Therefore, the significance of margins is a very controversial issue with respect to the treatment of DTs. In our case, it is unfortunate that evaluating whether the resection ends were negative was difficult. When the tumor is unresectable, inoperable, or recurrent, or the primary tumor is incompletely resected, postoperative radiotherapy is recommended. For patients with a positive postoperative incisional margin, similar negative results can be achieved after radiotherapy. Chemotherapy is also an option for patients who are not sensitive to radiotherapy, Chemotherapy may be performed according to the patient's condition, and includes doxorubicin, vinblastine, methotrexate, dacarbazine, sulindac and tamoxifen, which have been used in the treatment of DTs, but the therapeutic effect is largely different. As this case was found by physical examination, and there was no absolute contraindication to surgery, surgical resection was chosen. There was no recurrence during the 9-month follow-up.

Conclusion

Preoperative retroperitoneal DFs are rare and a correct diagnosis is a clinical challenge. The possibility of retroperitoneal DF should be considered when clinicians find isointensity on T1W imaging, hyperintensity on T2W imaging and progressive enhancement on MRI after peritoneal tumor, especially in patients with a surgical history.

Declarations

Consent to publish

The patient provided written informed consent for publication of this case report and accompanying images.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Funding

This work was supported by the Program for Training Capital Science and Technology Leading Talents [grant number Z181100006318003]. The funding body contributes to the design of the study and analysis of data.

Authors' Contributions

AL, XS collected data during the study. XW contributed to the study design. XW, WY, YN developed the first draft of the manuscript which was then reviewed and intensively revised by HW and ST. All authors read and approved the final manuscript.

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